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Case Report

Adenoid cystic carcinoma of the external auditory canal

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Abstract

Primary cancers of the external auditory canal (EAC) are rare and most are squamous cell carcinomas. We report the case of a 78-year-old man who visited our institution with a 5-month history of right-side intermittent otalgia and ear fullness. Otoscopic examination showed a bulging mass arising from the superior and posterior aspects of the right EAC, and incision biopsy confirmed the lesion as adenoid cystic carcinoma (ACC). Lateral temporal bone resection in conjunction with total parotidectomy and neck dissection was subsequently performed. Postoperative adjuvant radiotherapy was administered and no recurrence was noted at a 26-month follow-up. We review the medical literature on the topic and suggest that early diagnosis is still the best option for successful treatment of this neoplasm. ACC arising in the EAC must be removed using radical procedures to increase the chance of local control. Subsequent metastasis that tends to develop in the lungs and regional lymph nodes is best evaluated regularly using computed tomography examination.

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1. Introduction

Malignant tumors involving the external auditory canal (EAC) and temporal bone are exceedingly rare, and the most common type is squamous cell carcinoma.¹ The presence of adenoid cystic carcinoma (ACC) arising in the EAC is therefore infrequently encountered. ACC of the head or neck is usually found in the salivary glands, oral cavity, palate, nasal cavity, and nasopharynx.² Since 1894, there have been only 106 cases of ACC involving the EAC reported in the English literature. The natural history of ACC of the EAC is characterized by an indolent clinical course, which usually leads to a late diagnosis. The treatment goal includes complete surgical extirpation and a clear margin because of the high risk of repeat local recurrence. It is not uncommon for distant metastasis, mainly to the lungs, to occur over the course of many years.^{3,4}

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Because of the rarity of ACC of the EAC, most of the observations drawn from various reports lack detailed comparisons of pathological findings and long-term outcome follow-up. Here, we present our own case along with a review and discussion of the literature to date.

2. Case report

A 78-year-old man presented with a 5-month history of right-side intermittent otalgia and ear fullness. His past history was unremarkable. Otoscopic examination showed a bulging mass with an irregular surface arising in the right EAC (Fig. 1A). High-resolution computed tomography (CT) of the temporal bone showed an ill-defined, moderately enhanced, soft tissue mass protruding from the superior and anterior walls of the EAC (Fig. 1B), measuring 10 mm at the widest point, with adjacent bone erosion. Incision biopsy confirmed the diagnosis of ACC. Further information from magnetic resonance imaging (MRI) with gadolinium suggested that the superficial lobe of the parotid gland and the anterior portion of the temporal bone were also

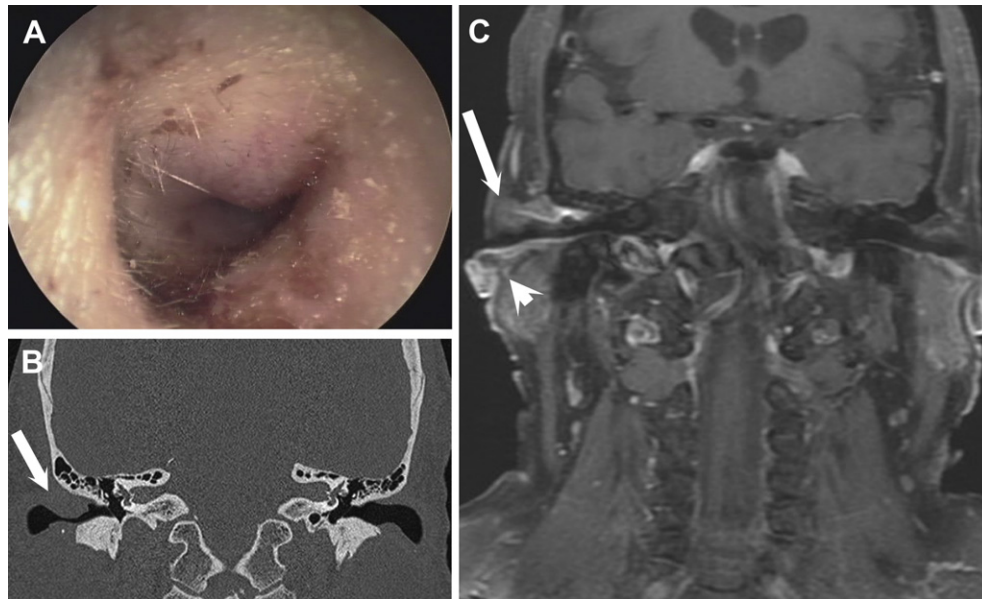


Fig. 1. (A) Subcutaneous mass bulging from the superior and posterior aspects of the external auditory canal (EAC). (B) Coronal computed tomography scan of the temporal bone demonstrating a soft tissue mass (arrow) protruding from the superior wall of the EAC. (C) Magnetic resonance imaging shows that the superficial lobe of the parotid gland (arrowhead) and the anterior portion of the temporal bone (arrow) may have been invaded by the mass lesion.

invaded (Fig. 1C). In view of the fact that the tumor had eroded the osseous EAC but had not invaded the middle ear cavity, and based on a presumed clinical T3N0M0 stage using University of Pittsburgh TNM staging for EAC carcinoma,^{5,6} surgery entailing lateral temporal bone resection in conjunction with total parotidectomy and selective neck dissection levels I–III was performed. In brief, the lateral temporal bone resection was approached with a wide postauricular incision followed by a separate incision made around the tragus and a portion of the conchal cartilage to isolate the tumor from the normal auricle. The facial nerve was skeletonized in its course through the stylomastoid foramen up to the geniculate ganglion and was found to be tumor-free. The malleus–incus joint was then disarticulated. The EAC, with the tumor lesion in its osseous and cartilaginous parts, was completely isolated and removed en bloc, along with the tympanic membrane and the ossicles (Fig. 2). The defect left over the lateral temporal region was reconstructed by rotation of the sternocleidomastoid (SCM) muscle and temporalis muscle flap.

Histologically, the tumor cells were in sheets and showed a predominantly cribriform pattern with scant intervening stroma. The cells were basaloid, with a scanty cytoplasm and hyperchromatic nuclei (Fig. 3). The lumina in the cribriform areas contained material that was positive in a periodic acid Schiff test. Few areas of necrosis were noted. Immunohistochemistry analysis revealed intense and diffuse positive staining for cytokeratin, vimentin, and muscle actin, and focal staining for S-100 protein. These findings confirmed the diagnosis of ACC of the EAC with perineural invasion and involvement of the tympanic portion of the temporal bone. The parotid gland and neck nodes were not invaded. Although clear surgical margins were achieved, postoperative adjuvant radiotherapy with doses of 6000 cGy in 34 fractions was administered. Good progress was noted during the patient's 26-month follow-up, and

the reconstructed external ear showed excellent esthetic and structural support (Fig. 4).

3. Discussion

ACC (also called cylindroma in the older literature), although it seldom arises in the EAC, is the most common malignant lesion of glandular origin.^{3,7,8} Our review of 106 cases of ACC arising from the EAC is based on the PubMed indexed English literature. Of the 106 ACC cases, 93 had clearly identified gender data, with a female-to-male ratio of 1.58 (57 vs. 36), indicating that females are more prone to being affected than males. Although ACC may appear at any age, the peak incidence is around the fourth and fifth decades of life. This tumor has an indolent clinical course and usually grows for years before causing symptoms, and thus tends to result in late diagnosis.

The most common clinical manifestations are otorrhea, pain, hearing loss, bleeding, and a mass or polyp in the ear canal.^{9–11} Differential diagnosis of a mass lesion in the EAC includes adenoma, papilloma, tuberculosis, and other neoplasms such as mucoepidermoid carcinoma, adenocarcinoma, basal cell carcinoma, and squamous cell carcinoma. Because of the propensity of ACC toward aggressive repeated local recurrences and distant metastases, it is important for clinicians to determine the extent of tumor invasion for adequate surgical planning and management.¹² The use of MRI in cases of ACC of the EAC has not been fully delineated.¹³ ACC occurring elsewhere in the head and neck may show low-signal intensity on T1-weighted (T1w) MRI images and high or low intensity on T2-weighted (T2w) images, depending on its cellularity. Our case presented with low intensity on T1w but high intensity on T2w images, indicating low cellularity, which was compatible with predominantly cribriform histological features.

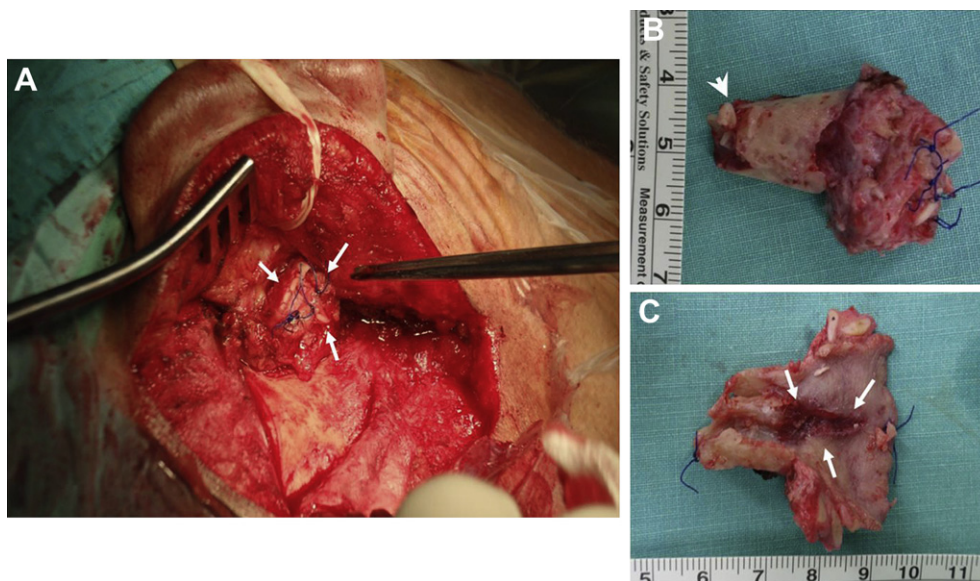


Fig. 2. Intraoperative photographs of the lateral temporal bone resection. (A) The operation started with obliteration of the orifice of the external auditory can (EAC; arrows) to prevent tumor seeding. (B) En bloc surgical removal of the lateral temporal bone along with the tympanic membrane and the ossicles (arrowhead). (C) The tumor origin was found in the superior and posterior aspects of the EAC (arrows).

The true origin of primary ACC in the EAC is still controversial, although it has been proposed that these tumors may arise from the eccrine sweat glands or ectopic salivary glands.¹⁴ Evidence of origin in the ceruminous glands has also been demonstrated in recent studies.⁷ Histologically, ACC can present in three patterns: tubular, cribriform, and solid.¹⁵ Immunohistochemical staining and electron microscopic examinations have revealed that the tubular pattern contains the greatest dual differentiation of the ceruminous glands (ductal and myoepithelial cells), while the solid pattern has the least. These findings more or less reflect the relationship between ACC and the ceruminous gland. In addition, it has been found that when arising from the salivary glands, these three ACC patterns show a significant correlation between histological pattern and prognosis,^{16,17} with the tubular pattern having the best prognosis. For ACC of the EAC, a correlation between histopathological status and clinical behavior has yet to be established. An analysis of the case presented here and 40 cases reported in the literature with clear histological descriptions revealed that the cribriform pattern has the

highest prevalence (27 cases, 65.9%), with tubular and solid features found in 11 (26.8%) and three (7.3%) cases, respectively (Table 1). However, the relationship between histopathology and prognosis remains obscure because of the limited number of cases and inadequate long-term follow-up. Nevertheless, it is believed that ACC arising in the EAC is far more aggressive than ACC in the salivary glands and leads to poor prognosis, in spite of the histological findings. Most authors have suggested that a solid histological pattern may be highly associated with the worst prognosis, as observed for ACC of the salivary glands. Further efforts involving detailed case studies and more reported cases will be needed to provide meaningful correlations.

ACC has a propensity towards early perineural involvement, which might explain why otalgia is one of the most common symptoms. In our review of the literature, the incidence of perineural involvement was estimated at 80% for 35 cases, including the present one, regardless of whether or not initial perineural invasion was clearly mentioned. Expression of neural cell adhesion molecules by neoplastic tumor cells

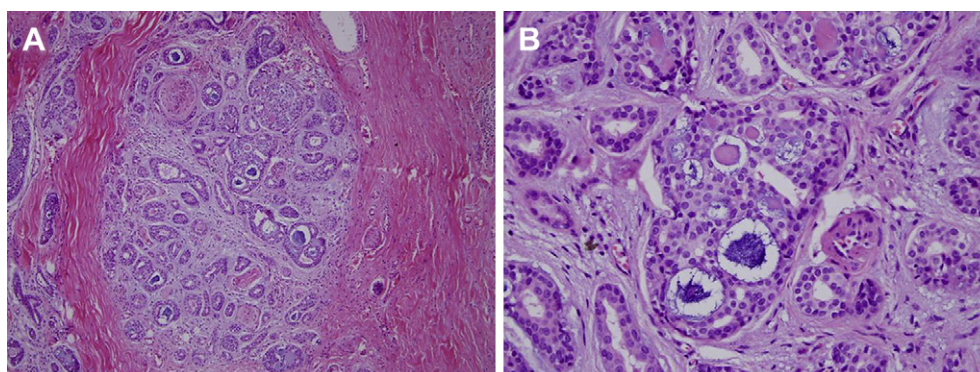


Fig. 3. (A) Micrograph of adenoid cystic carcinoma showing a characteristic cribriform pattern with perineural invasion (hematoxylin and eosin stain, 100×). (B) The lumina in the cribriform areas contained hyaline material (hematoxylin and eosin stain, 400×).



Fig. 4. The patient was monitored and was disease-free for 26 months. Excellent esthetic results were achieved.

has been proposed as an explanation for the perineural invasion of ACC that accounts for most intracranial involvement cases.¹⁸ When perineural invasion is identified, the tumor recurrence rate is higher (50% vs. 14.3%). Lymphovascular invasion is not common; our review revealed only four out of 35 cases (11.4%; Table 1). The tumor recurrence rate is also higher in lymphovascular invasion (66.7% vs. 23.1%).

In view of its locally aggressive behavior, treatment of ACC of the EAC depends on complete surgical resection with clear margins. In our case, the tumor was located just outside the tympanic annulus without invasion of the middle ear or mastoid cavities; therefore, lateral temporal bone resection was suitable as the surgical choice rather than more invasive subtotal

temporal bone resection or total temporal bone resection. Apart from surgical margins, involvement of the parotid gland, nerve, and bone are also important prognostic factors.¹⁹ Elective parotidectomy is recommended, even for patients in early ACC stages, because our review revealed an estimated 30% positive rate in patients with negative clinical parotid gland involvement preoperatively (12 out of 41 cases underwent parotidectomy). Metastases to regional lymph nodes and distant sites have been documented.^{3,20} In our review, occult lymph node metastasis was recognized in 11.1% (four out of 36) of cases who underwent neck dissection (Table 1).

Regarding the role of postoperative radiation therapy in ACC of the EAC, it is difficult to determine the impact of postoperative radiation on the outcome of a limited number of patients. However, on identifying specific tumor factors, Silverman et al suggested that advanced T-stage and positive microscopic margins are important factors in determining the necessity for postoperative radiation therapy for ACC of the head and neck.²¹ In addition, postoperative radiation is indicated for patients with ACC located in the sinonasal tract, cervical nodal metastasis, a predominant solid histologic pattern, perineural invasion, and recurrent tumors.^{22–25} For patients with early T-stage tumors and negative surgical margins, such as in our case, adjuvant radiation therapy may not be necessary.²¹ Use of radiotherapy alone yields a low survival rate.⁸ A recent study identifying the role of chemotherapy in salivary gland ACC showed that there is little evidence that systemic treatment for ACC alters its course.²⁶ Early diagnosis is still the best option for successful treatment.

In conclusion, ACC is a rare neoplasm originating in the EAC. The cribriform histological pattern is more predominant and demonstrates a high perineural invasion rate. Occult parotid gland involvement should not be overlooked. Delayed distant metastasis of this neoplasm to the lungs, kidneys, and vertebrae should be kept in mind. Most postoperative recurrences have been reported within 2 years following treatment, but the interval may extend to up to 15 years.³ Although the 5-year survival rate is high, owing to the high risk of local recurrence and delayed distant metastasis, a surgical approach

Table 1
Summary of the literature review of adenoid cystic carcinoma of the external auditory canal.

Reference	Patients	Histology (n = 41)			PNI (n = 35)	LVI (n = 35)	PARO (n = 41)	ND (n = 36)
		Tubular	Cribriform	Solid				
3	24	5	11	1	15/22	3/22	0/10	1/6
4	1		1					
8	1		1				0/1	0/1
9	2		1				0/1	
10	1	1			1/1	1/1	0/1	0/1
11	1		1					
12	1		1				0/1	0/1
13	1			1				
14	16	5	10	1	11/11	0/11	4/14	2/14
19	10						7/10	0/10
20	2						1/2	1/2
This case	1		1		1/1	0/1	0/1	0/1
Summary	61	11 (26.8%)	27 (65.9%)	3 (7.3%)	28 (80%)	4 (11.4%)	12 (29.3%)	4 (11.1%)

LVI = lymphovascular invasion; ND = neck dissection; PARO = parotidectomy; PNI = perineural invasion.

involving en bloc removal of the tumor with clear margins, followed by postoperative radiation and close long-term follow-up, is highly recommended.

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